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Case Report

Multimodality imaging of a cardiac paraganglioma

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ABSTRACT

Paragangliomas are rare neuroendocrine tumors arising from the ganglia of the sympathetic or parasympathetic nervous system. Less than 160 cases of intrapericardial or intracardiac paragangliomas have been reported in the English language peer-reviewed medical literature. Here, we report a case of intrapericardial paraganglioma, which illustrates some typical multimodality imaging features of this rare entity.

Case report

We report a case of a 25-year-old woman who was initially assessed with echocardiography for investigation of palpitations and mild exertional dyspnea. Transthoracic echocardiogram demonstrated a 6-cm mediastinal mass of heterogeneous echotexture in the right atrioventricular groove, compressing and deforming the right ventricle and right atrium (RA; Fig. 1).

The patient then underwent a non–EKG-gated computer tomography scan of the chest with intravenous contrast. It revealed a well-defined mass in the right atrioventricular groove, indenting the RA and right ventricular outflow tract, and abutting the aortic root and distal superior vena cava (Fig. 2). The mass was hyperenhancing with a thin hypoenhancing rim and a central area of stellate hypoenhancement, reminiscent of a central scar of hepatic fibronodular hyperplasia. The lesion contained neither detectable fat nor calcifications. It appeared intrapericardial and possibly intra-atrial.

The subsequent cardiac magnetic resonance imaging again demonstrated a well-defined right intrapericardial mass, showing avid enhancement with intravenous gadolinium contrast, and central stellate "scar" (Fig. 3). The appearance of the mass on cine images, and its broad base along the wall of the right cardiac chambers favored a mass centered on the epicardium.

(18)F-fluorodeoxyglucose positron emission tomography was also performed and showed a 6-cm intensely hypermetabolic lesion in the right heart with standardized uptake value of up to 31 and central necrosis (Fig. 4).

Coronary angiography was performed next and showed the mass's blood supply from a prominent branch of the proximal right coronary artery (Fig. 5). The mass enhanced rapidly and washed out quickly via a confluence of small veins, which drained into the coronary sinus.

An indium-111 octreotide single-photon emission computed tomography at 4 and 24 hours postadministration of the radiopharmaceutical was performed next. The lesion

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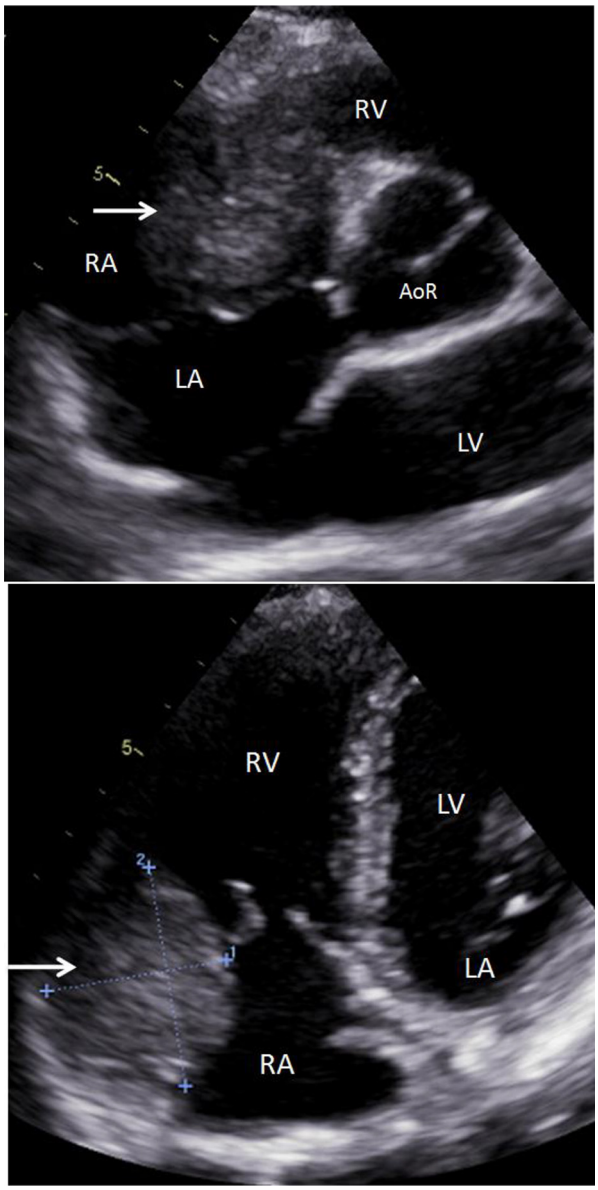

Fig. 1 – Transthoracic cardiac ultrasound demonstrating a heterogeneous mass (arrow) along the wall of the right heart chambers, at the level of the atrioventricular groove, abutting the aortic root, indenting and deforming the RA and RV. LA, left atrium; LV, left ventricle; RV, right ventricle; AoR, aortic root.

was positive for octreotide uptake (Fig. 6), thus confirming the suspected diagnosis of a cardiac paraganglioma.

An iodine-131 metaiodobenzylguanidine (I-131 MIBG) scintigraphy was also performed and showed mild heterogeneous radiotracer accumulation in the RA at the site of the mass (Fig. 7).

The patient eventually underwent surgical resection of the mass, with final pathology confirming the diagnosis of intrapericardial paraganglioma (IPP).

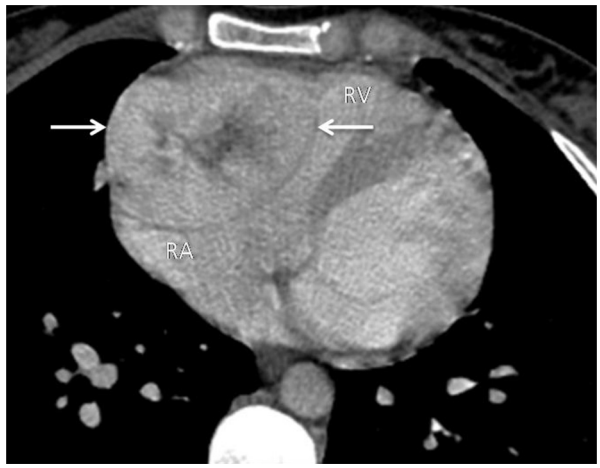

Fig. 2 – Contrast-enhanced computed tomography of the chest demonstrating a well-defined hyperenhancing mass (arrows) with a thin hypoenhancing rim, and a central area of stellate hypoenhancement, in the right atrioventricular groove, indenting the RA and right ventricle outflow tract.

Discussion

Paragangliomas are rare neuroendocrine tumors closely related to pheochromocytomas, tumors of chromaffin-positive cells of the adrenal glands [1]. Extra-adrenal paragangliomas may arise anywhere in the body, with the majority found in the abdomen [2]. Approximately 2% of paragangliomas occur in the thorax. IPPs are very rare. In 2014 Wang et al. [3] published a review article, where they identified 158 reported cases of cardiac paragangliomas in the English language PubMed database. They found male-to-female ratio of 0.86 and mean age at diagnosis of 39.7 years.

A minority of patients with IPP (about 8%) may be completely asymptomatic, and lesions might be found incidentally on imaging [4]. When present, symptoms in patients with IPP are usually secondary to excessive catecholamine excretion by the tumor, resulting in headaches, palpitations, diaphoresis, and systemic arterial hypertension [3].

On cross-sectional imaging, IPP usually appear as large, 3-8 cm, well-defined and often encapsulated heterogeneous masses [5]. Internal calcifications can be occasionally present [5]. IPP are hypervascular, demonstrating avid enhancement and rapid washout on contrast studies, and a hypoenhancing central area due to necrosis [6]. Most of the IPPs arise from the epicardium, and the majority are found in the interatrial and atrioventricular grooves or at the root of the great vessels, areas where parasympathetic ganglia normally exist [3].

The IPP are supplied by the coronary arteries, the majority (57.9%) fed by the right coronary artery [3].

At echocardiography, paragangliomas usually appear as echogenic masses and could be mistaken for myxomas, when arising from the interatrial septum [6]. Unlike myxomas, however, paragangliomas have a broad base [6]. Compression of adjacent vascular structures and encasement of the coronary arteries may be seen [6].